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Case Report

An unusual cause of pulmonary artery hypertension in a patient with chronic obstructive airway disease

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ABSTRACT

Pulmonary artery hypertension is a common entity in patients with obstructive airway disease mostly associated with chronic hypoxia. Presence of pulmonary artery hypertension can add to the respiratory distress patients experience during the clinical course of the disease. This is a case of a 55 year old man with severe chronic obstructive pulmonary disease with the alternative diagnosis of anomalous pulmonary venous return as a cause of pulmonary hypertension and persistent dyspnoea. This alternative etiology can lead to unsatisfactory results inspite of optimal treatment and therefore needs to be addressed individually.

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Educational aims

- To understand the alternative pathologies in patient with obstructive airway disease.
- Work up of pulmonary artery hypertension.
- To understand the pathology of atrial septal and pulmonary venous defects.
- Medical and surgical management of PAPVC.

1. Case report

A 55 year old African American male with a smoking history of 20 pack years had been complaining of progressively increasing shortness of breath with exertion. His medical history was significant for stroke in the past, benign prostatic hyperplasia and chronic renal insufficiency. His home medication consisted of terazosin and aspirin.

On physical examination lungs revealed bilateral decreased breath sounds with hyperinflation of the chest, heart sounds were difficult to appreciate in view of large lung volumes but a holosystolic murmur was heard best in the tricuspid area. There was jugular venous distention 2–3 cm above clavicular border, mildly tender hepatomegaly and one plus peripheral edema till ankles. Chest radiograph and tomogram showed cardiomegaly and central venous congestion (Figs. 1 and 2).

Patient was diagnosed to have chronic obstructive pulmonary disease when on pulmonary function testing there was a very severe obstruction with an insignificant bronchodilator response and an increased RV/TLC suggesting air trapping. DL_{CO} was also severely reduced. proBNP was 9289.

At the time of diagnosis of COPD patient's echocardiogram showed normal left ventricular size and wall thickness. Normal left ventricular function with all segments contracting normally. Left ventricular ejection fraction was ranging from 60 to 65%. Additionally there was a moderate right ventricular enlargement, moderate left atrial enlargement without thrombus and a marked right atrial enlargement. Calculated pulmonary artery systolic pressures of 73 mm Hg and a tricuspid regurgitation jet of 3.8. A V/Q lung scan was negative for any pulmonary thromboembolism.

Patient was subsequently setup for a right heart catheterization which revealed pulmonary artery systolic pressure of 70 mm Hg, pulmonary artery diastolic pressure of 25 mm Hg with a mean pulmonary artery pressure of 40 mm Hg. Right atrial pressure was 8 mm Hg with a pulmonary capillary wedge pressure of 14 mm Hg. Cardiac output could not be obtained due to severe tricuspid regurgitation. Oxygenation at different sections of the right side of the heart was SVC: PaO_2 39 mm Hg SaO_2 71.5%. RA:

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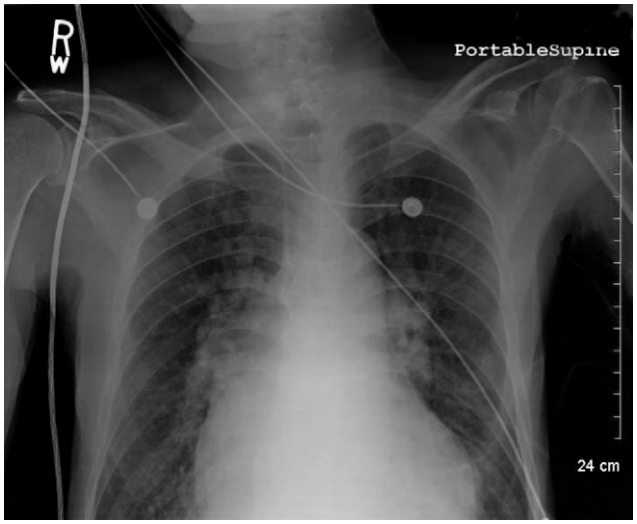

Fig. 1. Chest X-ray show bilateral hyperinflated lung, cardiomegaly and perihilar prominence signifying pulmonary artery hypertension and right sided enlargement of the heart.

PaO_2 61 mm Hg SaO_2 92%. PA: PaO_2 91 mm Hg SaO_2 97%. Thus there was a step up from SVC to RA to PA in oxygen saturation, indicating L to R shunt.

A Contrast study done by a TTE (trans-thoracic echocardiogram), in order to assess for a thin foramen ovale as the possible cause of pulmonary artery hypertension, showed immediate opacification of 50–60% of the left atrium without an obvious patent foramen ovale or secundum atrial septal defect but was suggestive of a sinus venosus atrial septal defect. To further investigate this a TEE (Trans-esophageal echocardiogram) was done which showed sinus venosus defect with incorporation of the right upper pulmonary vein into the sinus venosus.

2. Discussion

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital anomaly where one or more but not all pulmonary veins fail to connect to the left atrium. It is present in only 0.4–0.7% of postmortem examinations. These anomalous pulmonary veins usually occur in the right lung with only 10% originating from the left lung.[1]

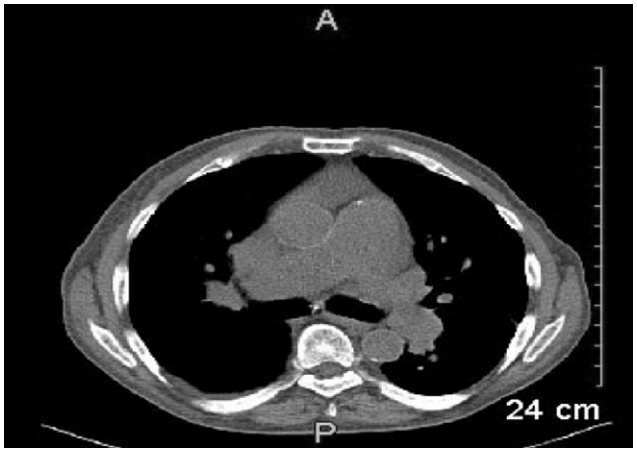

Fig. 2. Chest tomogram of the patient shows enlarged pulmonary artery in comparison to the aorta and small right sided pleural effusion.

PAPVC often presents with few clinical symptoms and may remain undiagnosed until late adulthood. When associated with atrial septal defect (ASD), however, patient may have significant symptoms and present in early childhood due to increase in left to right shunting. About 10% of patients with atrial septal defect have one or more abnormally draining pulmonary veins.[2]

PAPVC of three types may be suspected with an intact atrial septum. The most common variety is the anomalous drainage of the right upper and middle lobe pulmonary veins to the right atrium or superior vena cava (about 90%) as demonstrated in our patient. Drainage of the left pulmonary veins to the innominate vein via an abnormal venous channel is the next common variety. Rarely a patient may have left pulmonary veins draining into the coronary sinus.

As mentioned before, most patients with PAPVC are asymptomatic. If a significant left-right-shunt coexists, patient may develop irreversible pulmonary hypertension, pulmonary vascular obstructive disease, or right ventricular failure.[3] Pulmonary hypertension could be a late manifestation of isolated partial anomalous venous drainage occurring due to increased pulmonary blood flow leading to reflex pulmonary vasoconstriction and eventually pulmonary vascular obstructive disease.[4] The presentation varies from dyspnoea and fatigue with exertion to no symptoms early in the life. A soft systolic murmur due to increased blood flow across the pulmonary valve along with fixed splitting of the second heart sound during respiration is present on examination.

The early diagnosis of the defect can be made by echocardiography especially trans-esophageal echocardiography that has replaced cardiac catheterization as the gold standard.[4,5]The presence of tricuspid regurgitation on echocardiography may be a surrogate of the early stages of the right ventricular overload and should be considered a marker that pulmonary vascular disease has occurred. The role of fetal echocardiography in the prenatal diagnosis of total or partial anomalous pulmonary venous connection has also been approved in a Canadian study.[6] Surgical treatment should be offered to patients of PAPVC with evidence of right ventricular dilatation, mild to moderate tricuspid regurgitation, or early stages of pulmonary vascular disease in order to prevent the progression of right ventricular failure and irreversible pulmonary vascular disease. A study done by ElBardissi et al., proved the role of anterolateral thoracotomy without the use of cardio-pulmonary bypass resulting in excellent long-term outcome when performed correctly.[1]

In this patient because of his previous stroke and chronic renal insufficiency he was not a good candidate for surgical closure of his sinus venosus atrial septal defect. A decision regarding trial of medical therapy was made subsequently in accordance with patient's wishes. Patient initially was started on sildenafil at a dose of 20 mg three times a day. Since patient was a high risk patient with proBNP >4000, WHO functional class 3 symptoms, right ventricle dilation per echocardiogram, medication was changed to ambrisentan and then bosentan but was intolerant to both. Patient refused inhaled iloprost because of logistic problems but agreed to a non-conventional monotherapy with intravenous prostanoid therapy which was started at a dose of 1.25 ng/kg/min. The dose was slowly titrated up to 30 ng/kg/min with which patient showed improvement in his symptoms.

Conflict of interest statement

This is to state that no conflict of interest exists amongst any of the authors of the case report tilted: An unusual cause of pulmonary artery hypertension in a patient with chronic obstructive airway disease.

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